CLINICAL NOTES IN DIAGNOSTIC CARDIOLOGY

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Hypertrophic Cardiomyopathy and Septal Myectomy

JOHN S. CHILD, MD
WILLIAM R. CABEEN, JR., MD
NIGEL K. ROBERTS, MD
Los Angeles

HYPERTROPHIC CARDIOMYOPATHY (also known as idiopathic hypertrophic subaortic stenosis) is an uncommon genetically transmitted (non-sex linked, autosomal dominant) cardiomyopathy characterized by pronounced left ventricular hypertrophy, particularly of the interventricular septum. This disease covers a spectrum, with one end being characterized by systolic displacement of the anterior mitral leaflet into the left ventricular outflow tract into apposition with the ventricular septum, resulting in dynamic left ventricular outflow tract obstruction and mitral regurgitation. Anatomic characteristics include (1) asymmetrical septal hypertrophy and (2) histologic septal (sometimes free wall) myofibrillar disorganization.

This uncommon, but by no means rare, process is of import to general medical practitioners because it may be confused with aortic valvular stenosis or coronary artery disease because of symptoms of exertional dyspnea or angina pectoris or because of abnormalities shown on electrocardiograms. Echocardiography may assist in diagnosis by showing asymmetric septal hypertrophy and, in those patients with left ventricular outflow tract obstruction, systolic anterior motion of the mitral valve.[1-3] Awareness of the safety and ability of echocardiography in diagnosis of hypertrophic cardiomyopathy can assist physicians in decisions regarding more invasive procedures (cardiac catheterization) and in institution of appropriate therapy.

A case is presented that exemplifies many of the characteristics of hypertrophic cardiomyopathy.

Report of a Case

A 34-year-old white man was well until 1974 when exertional dyspnea and syncope during exercise developed. He sought cardiologic advice. An echocardiogram and a cardiac catheterization were reported to show idiopathic hypertrophic subaortic stenosis. Propranolol (Inderal) therapy resulted in significant improvement but was discontinued in 1975 because of possible adverse psychiatric effects. The exertional dyspnea increased, and a precordial "pressure" sensation began to occur during exertion, relieved by rest and administration of nitroglycerin. An electrocardiogram during pain showed ST segment depression. Therapy with propranolol (40 mg given orally four times a day) was again instituted but the patient's symptoms did not greatly decrease. He discontinued his work in a mechanical parts shop. Because of increasing angina (occurring once or twice a day) with minimal exertion, as well as a near-syncopal episode, he was referred to the University of California, Los Angeles, Hospital in 1976. Family history showed that the patient's father had died suddenly at the age of 42 and had had no history of cardiac difficulties. It also was noted that a 28-year-old sister of the patient has a heart murmur (cause unknown).

On physical examination, blood pressure was 120/70 mm of mercury, pulse rate was 70 beats per minute (regular), no abnormal jugular venous distention or waves were noted and there was a double peaked carotid impulse (with normal upstroke and volume). The left ventricular apical impulse was palpable in the fifth left intercostal space just beyond the midclavicular line and was single. The first (S_1) and second (S_2) sounds were normal, but there was an atrial filling gallop audible at the apex. A grade III/VI high pitched crescendo-decrescendo (ejection) systolic murmur was heard at the apex and along the left sternal border, which began just after S_1 and ended before S_2. The murmur did not change with

From the Departments of Medicine and Pediatrics, Division of Cardiology, University of California, Los Angeles, Center for the Health Sciences.

Reprint requests to: John S. Child, MD, Division of Cardiology, UCLA Department of Medicine, 10833 Le Conte Avenue, Los Angeles, CA 90024.

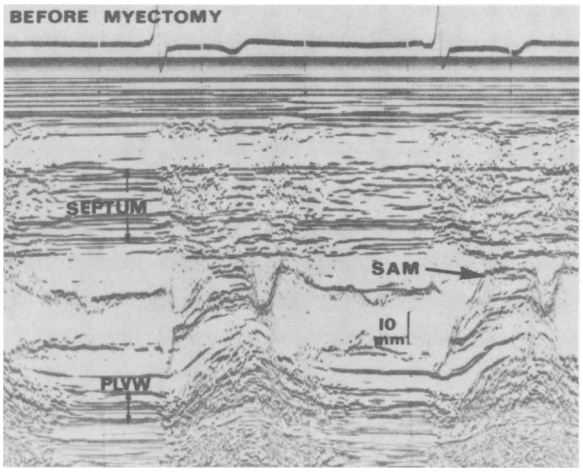

Figure 1.—Preoperative echocardiogram of mitral valve and left ventricle. Shown are the mitral valve with systolic anterior motion (SAM) and asymmetric septal hypertrophy (septum/posterior left ventricular wall [PLVW] ratio = 2.4). These findings are characteristic of hypertrophic cardiomyopathy with obstruction. Shown is a 10 mm calibration.

a Valsalva maneuver. No ejection click or diastolic murmur was detected.

A roentgenogram of the chest showed no abnormalities. An electrocardiogram showed left ventricular hypertrophy (voltage) and nonspecific ST-T abnormalities (strain). A standard M-mode echocardiogram gave the following findings: (1) asymmetric septal hypertrophy with a septal diastolic thickness of 24 mm and a posterior left ventricular wall diastolic thickness of 10 mm, with a resultant intraventricular septum/left ventricular wall ratio of 2.4; measurements were taken where the ultrasonic beam traversed the mid-left ventricle, at the tip of the anterior mitral leaflet, and at the mid-portion of the posterior mitral leaflet (Figure 1); (2) systolic anterior motion of the mitral valve consistent with left ventricular outflow tract obstruction (Figure 1); (3) a hypokinetic ventricular septum with normal posterior left ventricular wall motion and systolic thickening; (4) normal-to-small left ventricular diastolic dimension of 36 mm, taken at the peak of the R-wave, at a beam angle just below the tip of the mitral valve (not shown in the figure); (5) normal end-systolic size of the left atrium (36 mm) and of the aortic root (26 mm), and (6) midsystolic notching of the aortic valve (Figure 2), consistent with dynamic left ventricular outflow tract obstruction and reflecting the physical and phonocardiographic finding of a bifid carotid impulse.

Cardiac catheterization (rest, supine) showed a resting peak-to-peak gradient of 60 to 70 mm of mercury across the left ventricular outflow tract which notably increased in the postpremature ventricular contraction beat in association with a slight decrease in aortic pulse pressure.

Despite increases in propranolol dosage over six weeks to 400 mg per day, the symptoms and echocardiographic findings were unchanged. Consequently, a septal myotomy-myectomy was carried out. Postoperatively, there was a left bundle branch block. A postmyectomy echocardiogram showed a decrease in systolic anterior motion of the mitral valve, and a decrease in septal diastolic thickness to 17 mm (Figure 3).

Although there has been some decrease in symptoms in the patient, continued propranolol therapy has been necessary to provide adequate control.

Discussion

Hypertrophic cardiomyopathy may have no demonstrable left ventricular outflow tract ob-

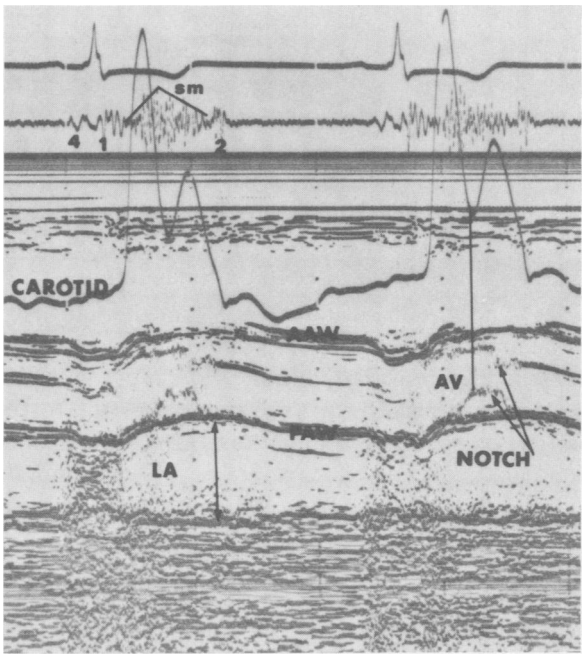

Figure 2.—Preoperative echocardiogram of aortic valve. Note the systolic notch of the aortic valve (AV) consistent with dynamic left ventricular outflow tract obstruction. This nicely correlates with the bifid carotid pulse tracing. The left atrium (LA) is normal in size. The phonocardiogram trace (from left lower sternal border) at the top of the figure shows the first (S_1) and second (S_2) heart sounds, an atrial filling gallop (S_4) and a crescendo-decrescendo murmur (outlined with solid lines). AAW = anterior aortic wall. PAW = posterior aortic wall.

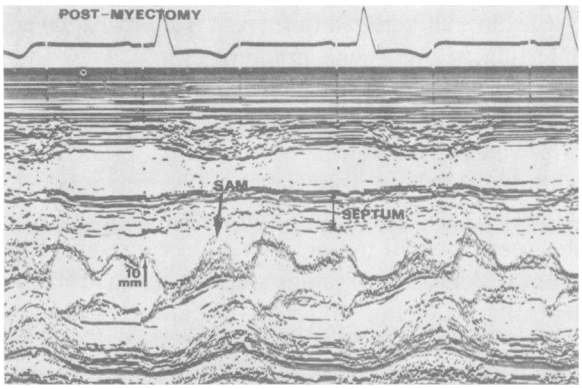

Figure 3.—Postmyectomy echocardiogram of left ventricle and mitral valve. The septum (17 mm) is now 7 mm thinner than the preoperative measurement (24 mm). Systolic anterior motion (SAM) is smaller in amplitude and duration than preoperatively, suggesting a lesser degree of outflow tract obstruction. The electrocardiographic lead now reveals an intraventricular conduction defect. Shown is a 10 mm calibration.

struction, or may have basal or provocable obstruction. Obstruction is due to systolic "tethering" of the anterior mitral leaflet into the left ventricular outflow tract and into apposition to the hypertrophied ventricular septum. The anterior mitral leaflet and upper ventricular septum in such patients show fibrous contact plaques pathologically. On physical examination, evidence for obstruction in the bifid carotid pulse, often a triple apex impulse, and a murmur are shown. Mitral regurgitation occurs in obstructive cardiomyopathy because of the abnormal systolic mitral valve motion. Such an abnormality renders these patients susceptible to bacterial endocarditis.

It is important to understand that the left ventricular outflow tract obstruction by the hypertrophied septum and mitral valve is a dynamic process governed by the following: (1) the force of left ventricular contraction; (2) the systolic left ventricular volume, and (3) the transmural distending pressure of the left ventricular outflow tract. Vasoactive drugs, such as digitalis glycosides or isoproterenol (increased inotropy) or nitroglycerin (decreased venous return), can increase obstruction and should be used with caution.

The postpremature ventricular contraction beat generates increased contractility and pressures in normal persons, in persons with fixed aortic stenosis and in persons with dynamic left ventricular outflow tract obstruction (hypertrophic cardiomyopathy). However, hypertrophic cardiomyopathy differs not in that the postpremature ventricular contraction gradient increases, but in that the postpremature ventricular contraction aortic pulse pressure is no greater than the prepremature ventricular contraction control.

Because exercise results in increased contractility, left ventricular outflow tract obstruction increases. Often as important is the immediate postexercise period when venous return decreases while sympathetic tone is still increased, therefore resulting in even higher gradients.

The Valsalva maneuver, or assuming the upright posture (squat to standing), decreases venous return, often causing an increase in obstruction, and on physical examination an increase in the intensity of the systolic murmur is noted. Similarly, hypovolemia may be poorly tolerated, as in surgical operations or parturition.

Just as important as left ventricular outflow tract obstruction is the restriction to left ventricular filling by decreased left ventricular compliance due, at least in part, to massive hypertrophy.[4,5] This results in an increase in left ventricular end-diastolic pressure and an increase in mean left atrial pressure. Physical examination, therefore, often shows an S_4 gallop. An increase in left atrial pressure and size predisposes to atrial fibrillation. This rhythm may be tolerated poorly by a stiff left ventricle because of the fast irregular ventricular rate and the decrease in available diastolic filling time. Also, the loss of the atrial "kick" may be important. Atrial fibrillation may be a medical emergency, and control of the heart rate is crucial, whether with propranolol or digoxin. Because systemic embolism is frequent, anticoagulation is necessary in the presence of atrial fibrillation.

Exertional dyspnea, as in our patient, is probably due to the left ventricular inflow obstruction and resultant increases in pulmonary venous pressure. The explanation for chest pain is less clear. The pain is often classic for angina pectoris, but the extramural coronary arteries are usually normal. Syncope and presyncope are often unrelated to the presence or absence of left ventricular outflow tract obstruction. Ventricular arrhythmias are accountable in some instances.

Sudden death is a major problem and is undoubtedly due in many instances to tachycardia or bradycardia. Worsened left ventricular outflow obstruction may also be contributory.

Because the symptoms described also may result from other diseases, echocardiography may be useful; if it gives the findings noted in the case discussed, a diagnosis of typical hypertrophic

cardiomyopathy with obstruction can be made (Figures 1 and 2). The dominant finding for hypertrophic cardiomyopathy (with or without obstruction) is asymmetric septal hypertrophy with a minimum septal/posterior left ventricular wall ratio of 1.3.[1] Integral to this criterion is that the septum is in fact thick (that is, thicker than 12 mm). Use of a higher septal/posterior left ventricular wall ratio (1.5 or more) is often recommended, and is preferred in our laboratory.[2] The septum is usually hypokinetic; that is, it usually has reduced systolic thickening and excursion.[3] In patients with left ventricular outflow tract obstruction, systolic anterior motion of the anterior mitral leaflet is also present, and usually midsystolic notching of the aortic valve.[3] The mitral valve typically touches the septum in diastole and often has a reduced diastolic closure rate. The left ventricular dimensions are almost always small-to-normal, and are essentially never increased. In addition, left atrial enlargement, with its propensity for atrial fibrillation, may be detected.

Treatment first should be directed at the significant risk of sudden death. Therefore, strenuous activity should be advised against to avoid inducing or increasing left ventricular outflow tract obstruction. Use of digitalis glycosides, nitroglycerin, isoproterenol or vigorous diuresis (or other causes of hypovolemia) may increase left ventricular outflow tract obstruction. Infective endocarditis prophylaxis should be recommended.

Propranolol (a beta-blocker) usually decreases left ventricular outflow tract obstruction due to tachycardia and sympathetic stimulation (such as exercise). It may also be useful in control of ventricular and atrial arrhythmias.

In the presence of pronounced basal left ventricular outflow tract obstruction, and disabling angina or syncope uncontrolled by propranolol therapy, with no alternative explanation for these symptoms, surgical operation may be indicated.[6] In the case discussed, septal myotomy-myectomy was carried out and there was some relief of symptoms. Of interest is the postoperative echocardiogram in this case (Figure 3) showing a thinner ventricular septum, and a decrease in systolic anterior motion of the mitral valve (suggesting a decrease in outflow obstruction). The only complication in the patient was left bundle branch block, a common finding. Other potential complications include ventricular septal defect and increased mitral regurgitation. Operative mortality in experienced hands is 5 percent.[7]

Prognosis appears to be worse with increased age, lack of obstruction, a high left ventricular end-diastolic pressure and, possibly, atrial fibrillation.[6] In general, those patients who are asymptomatic do well; those with symptoms have progressive difficulty. Surgical therapy seems to result in significant improvement in a large number of selected patients.[7]

Surgical therapy does not, however, have any clear-cut effect on the incidence of sudden death. Unfortunately, there are no clear clinical indicators that would help predict sudden death; one would expect, however, that detection by standard electrocardiography or Holter monitoring of significant arrhythmias may indicate a subgroup of patients at risk.

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